

. 论 著 .

颅颈交界区硬脊膜动静脉瘘的诊治：
附 2 例报道并文献复习

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【摘要】目的 探讨颅颈交界区硬脊膜动静脉瘘的诊断和治疗。方法 回顾性分析 2 例颅颈交界区硬脊膜动静脉瘘的临床资料,对其诊断、治疗方案及结果进行分析,并结合相关文献进行总结。结果 2 例均行选择性脑脊髓动脉造影术确诊,均行显微手术成功灼闭瘘口,术后感觉障碍减轻、四肢肌力及大小便功能正常。结论 选择性脑脊髓动脉造影术是诊断颅颈交界区硬脊膜动静脉瘘的金标准,手术灼闭瘘口是治疗颅颈交界区硬脊膜动静脉瘘的有效治疗方案。

【关键词】硬脊膜动静脉瘘;颅颈交界区;选择性脑脊髓动脉造影;显微手术

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Diagnosis and treatment of spinal dural arteriovenous fistulas at the craniocervical junction: two case reports and literature review

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【Abstract】Objective To explore the diagnostic and therapeutic methods for spinal dural arteriovenous fistulas (SDAVFs) at the cranio-cervical junction. Methods The clinical data of two patients with SDAVFs at the cranio-cervical junction were retrospectively analyzed. The diagnosis, treatment plans, and outcomes were elaborated in detail, and a summary was made in combination with the relevant literature. Results Both patients were diagnosed by selective cerebral and spinal angiography and successfully underwent microsurgical cauterization of the fistula. Postoperatively, their symptoms improved significantly, sensory disturbances were mitigated, and muscle strength in the limbs as well as bladder and bowel functions returned to normal. Conclusion Selective cerebral and spinal angiography is the gold standard for diagnosing SDAVFs at the cranio-cervical junction, and microsurgical cauterization of the fistula is an effective treatment approach.

【Key words】Spinal dural arteriovenous fistula; Craniocervical junction; Selective cerebral and spinal angiography; Surgery

脊髓硬脊膜动静脉瘘(spinal dural arteriovenous fistula, SDAVF)是一种常见的脊髓血管畸形,好发于胸腰段,颅颈交界区罕见^[1-3]。颅颈交界区 SDAVF (craniocervical junction SDAVF, CCJ-SDAVF)是发生于枕骨大孔区至第 2 颈椎之间的罕见脊髓血管疾病,约占所有 SDAVF 的 2%。CCJ-SDAVF 的临床表现多种多样,可无症状,也可出现蛛网膜下腔出血、脑干功能障碍或进行性脊髓病,应及早诊治^[4-7]。因为 CCJ-SDAVF 的发病率较低,而且此区域解剖结构复杂,临床表现各异,所以误诊率、漏诊率较高,从而影响病人的治疗和预后^[8]。本文总结 2 例 CCJ-SDAVF 的诊治经验,并进行文献分析,以提高对该疾病的认识。

1 病例资料

病例 1:49 岁男性,因颈痛伴四肢麻木、乏力 2 个月余于 2023 年 2 月 11 日入院。病程中出现会阴部感觉异常,偶有小便失禁、便秘。既往有高血压病史,未规律药物治疗及监测。入院体格检查:神志清楚,言语流畅;双侧瞳孔等大等圆,对光反射灵敏,双眼可见水平眼震;伸舌左偏;右上肢浅感觉稍减退,右上肱二头肌腱反射减弱;右侧胸 8 水平以下浅感觉减退,左侧胸 10 水平以下浅感觉减退;双上肢肌力 5 级,双下肢肌力 4⁺级。入院后行颈段 MRI 示延髓、颈段及上胸段脊髓广泛异常信号,颅颈交界区迂曲血管影(图 1A)。体感诱发电位(Somatosensory evoked potentials, SEP)及脑干听觉诱发电位(brainstem auditory evoked potentials, BAEP)示左侧大脑半球 SEP 异常(结合临床考虑右侧颈髓病变),右侧大脑半球 SEP 大致正常;双侧下肢 SEP 轻度异常(结合临床考虑脊髓病变);双侧 BAEP 轻度异常

(周围神经病变,右侧稍重)。行头颈部 320-CTA 示延髓、颈段血管异常,动静脉瘘可能(图 1C~E)。全脑血管造影术发现左侧颈外动脉分支咽升动脉供血的动静脉瘘(图 1F)。术前将 3D-DSA 与颅骨图像融合并结合 320-CTA 图像,判读瘘口与枕骨大孔、寰枢椎及脊髓关系,规划手术入路。在电生理监测下,采用枕下后正中入路,暴露枕骨、枕骨大孔及寰枢椎,去除寰椎后弓及部分枕骨,切开硬膜探查脊髓表面迂曲扩张血管并发现一异常血管穿透硬膜与其沟通,吲哚菁绿荧光造影确认动静脉瘘供血动脉后,临时动脉瘤夹夹闭复查荧光造影确定瘘口消失,解除临时动脉瘤夹,电凝灼闭瘘口后见脊髓表面原迂曲扩张血管颜色逐渐变暗淡,术后颈部疼痛感逐渐减轻(图 1G、1H)。术后 3 个月电话随访,颈痛及四肢麻木感消失,改良 Rankin 量表(modified Rankin scale,mRS)评分 0 分。

病例 2:61 岁男性,因突发头痛伴恶心呕吐 1 d 于 2023 年 6 月 11 日入院。既往有高血压病史,未规律服药。入院体格检查:神志清楚,颅神经未见明显异常;左侧肢体肌力 5 级,右侧肢体肌力正常,四肢肌张力正常。结合病史、临床表现、出血部位,考虑为高血压性脑出血可能(图 1A),完善全脑血管造影术排查时发现 CCJ-SDAVF,由左侧椎动脉脑膜分支参与供血,引流静脉位于脑干腹侧,向同侧岩下窦方向引流(图 2B~D)。术前将 3D-DSA 与颅骨图像融合,判读瘘口与枕骨大孔、寰枢椎及脊髓关系,规划手术入路。在电生理监测下,采用左侧远外侧入路,暴露枕骨、枕骨大孔及寰枢椎,去除寰椎后弓及部分枕骨,切开硬膜后沿脑干外侧暴露椎动脉,探查见延髓表面及腹侧迂曲血管影及一粗大、动脉化的搏动明显的引流静脉向幕上方向引流,沿椎动脉临时夹闭阻断瘘口后见远端引流静脉张力下降,颜色渐变为暗黑色静脉,解除阻断后电凝灼闭瘘口(图 2E)。术后 2 周复查全脑血管造影显示瘘口消失(图 2F)。术后 3 个月电话随访,无特殊不适,mRS 评分 0 分。

2 讨论

CCJ-SDAVF 以中老年人常见,男性多见(男/女约 6:1)。与其他部位 SDAVF 有所差异,CCJ-SDAVF 的临床表现多种多样^[9-12],包括自发性蛛网膜下腔出血、慢性进展性脊髓病、脊髓内出血、脑干功能障碍和脊髓神经根疾病。本文 2 例中,1 例因右侧基底节区高血压性脑出血后行脑血管造影检查发现,另外 1 例因脑干脊髓功能障碍行脑脊髓血管造影确诊。

SDAVF 常与外伤、感染性静脉炎、凝血功能障碍、手术等因素相关^[2]。目前,其发病机制尚不明确,推测:硬脊膜静脉窦内血栓形成引发静脉压力增高而影响正常静脉回流,从而激活血管生长因子或毛细血管通路开放形成,脊髓冠状静脉、根静脉内压力升高,髓内动静脉压力梯度下降,脊髓静脉充血、静脉血管迂曲扩张,导致脊髓灌注下降、脊髓缺血^[12]。

由于 CCJ-SDAVF 起病隐匿,早期诊断困难。临床高度怀疑时,需进一步行影像学检查,临床上常常首选 MRI 筛查,其 T₂ 像可有脊髓高信号及脊髓周围异常血管流空影表象。必要时行 CTA 检查,在此基础上进一步行选择性脑脊髓动脉造影明确诊断,能清楚显示供血动脉、瘘口部位和引流静脉,而且,选择性脑脊髓动脉造影是诊断的金标准。CCJ-SDAVF 通常由椎动脉、枕动脉、咽升动脉、脊髓前动脉的脑膜分支动脉和硬脊膜根动脉参与供血,主要向髓静脉、冠状静脉丛、颅内静脉系统、脊髓前静脉和硬脊膜外静脉丛回流,静脉回流向颅内时,可因静脉内压力增高而引起蛛网膜下腔出血。当静脉流量不高时,颅颈交界区的逆行上行静脉引流可能诱发脑干静脉充血水肿。文献报道,出血的危险因素包括颅内静脉系统回流、静脉湖扩张、供血动脉血流相关性动脉瘤以及脊髓前动脉分支参与供血^[6,9,13]。

CCJ-SDAVF 主要采用显微手术、血管内介入栓塞治疗和复合手术治疗,治疗的目的是阻断异常的动静脉分流以缓解静脉充血,并尽可能保持正常的静脉引流。目前,显微手术为首选治疗方案,临床预后更好、治愈率高^[13-15]。手术方案的选择主要依据于动静脉瘘的具体血管构筑情况制定个体化治疗方案。对于血管构筑相对简单的动静脉瘘,像瘘口位置、引流静脉位于脊髓被侧或外侧且脊髓动脉分支不参与供血者,可采取简单的显微手术治疗,关键是灼闭、切断靠近瘘口附近的供血动脉及引流静脉,并且手术全程在神经电生理监测下进行,以便于保护、监测脊髓神经功能,同时术中采取闭塞试验、吲哚菁绿荧光造影来帮助定位和识别供血动脉、瘘口及引流静脉。条件允许情况下,可采取复合手术。术中造影进一步辨别,以便完全灼闭切断瘘口供血,达到治愈疾病的目标。然而,对于合并有脊髓动脉分支参与供血或者病变瘘口位于脊髓腹侧时,其血管构筑更加复杂多变,且部分处于出血急性期,显微手术灼闭瘘口、引流静脉或者夹闭脊髓动脉瘤样结构时较为困难,则可考虑先行血管内介入治疗,临床有时需要分次栓塞。血管内介入栓塞治疗的关键在于使

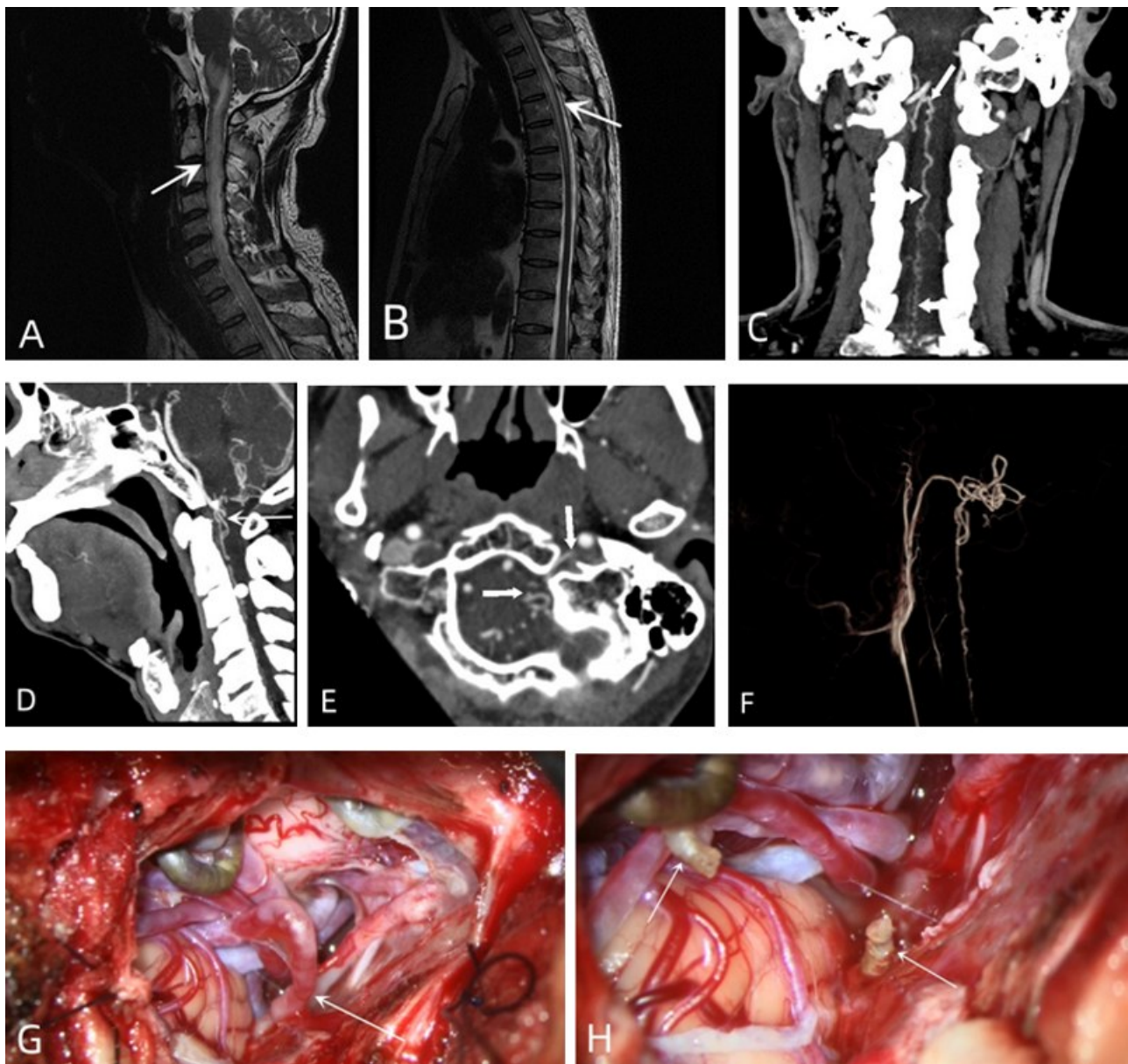


图1 颅颈交界区硬脊膜动静脉瘘的术前影像及术中显微镜下表现

A、B. 术前颈椎、胸椎MRI平扫，↑示延髓、颈段及上胸段脊髓异常信号；C~E. 术前头颅320-CTA示颅颈交界区动静脉瘘，↑示异常血管显影；F. 左侧颈外造影显示动静脉瘘供血及回流静脉；G、H. 术中显微镜下观察动静脉瘘，↑示供血动脉

Figure 1 Preoperative images and intraoperative microscopic manifestations of a patient with spinal dural arteriovenous fistula at the craniocervical junction

A-B: Preoperative cervical and thoracic MRI plain scans, with arrows indicating abnormal signals in the medulla oblongata, cervical, and upper thoracic spinal cord. C-E: Preoperative 320-CTA of the head reveals arteriovenous fistulas at the craniocervical junction, with arrows indicating abnormal vascular imaging. F: Left external carotid artery angiography shows the feeding artery and reflux vein of the arteriovenous fistula. G-H: Intraoperative microscopic observation of the arteriovenous fistula, with arrows indicating the feeding artery.

用栓塞材料完全闭塞瘘口，尤其是显微手术中难以暴露处理的部位，同时需将引流静脉起始端闭塞部分以降低术后复发风险，微导管顺利到位后，需多角度及超选择造影评估供血动脉、引流静脉的血流方向，明确微导管与瘘口的关系。根据造影剂弥散速度评估栓塞材料能否安全到达瘘口，从而闭塞供血

动脉、瘘口及引流静脉起始部分，常见的栓塞材料有NBCA胶、Onyx胶、弹簧圈、丝线线段等，在血管内介入栓塞后再考虑行二期显微手术治疗。有学者在显微手术过程中同时采取显微镜及神经内镜双镜，以便达到一次性治愈的目的。

CCJ-SDAVF临床表现多样，自然转归不良，且

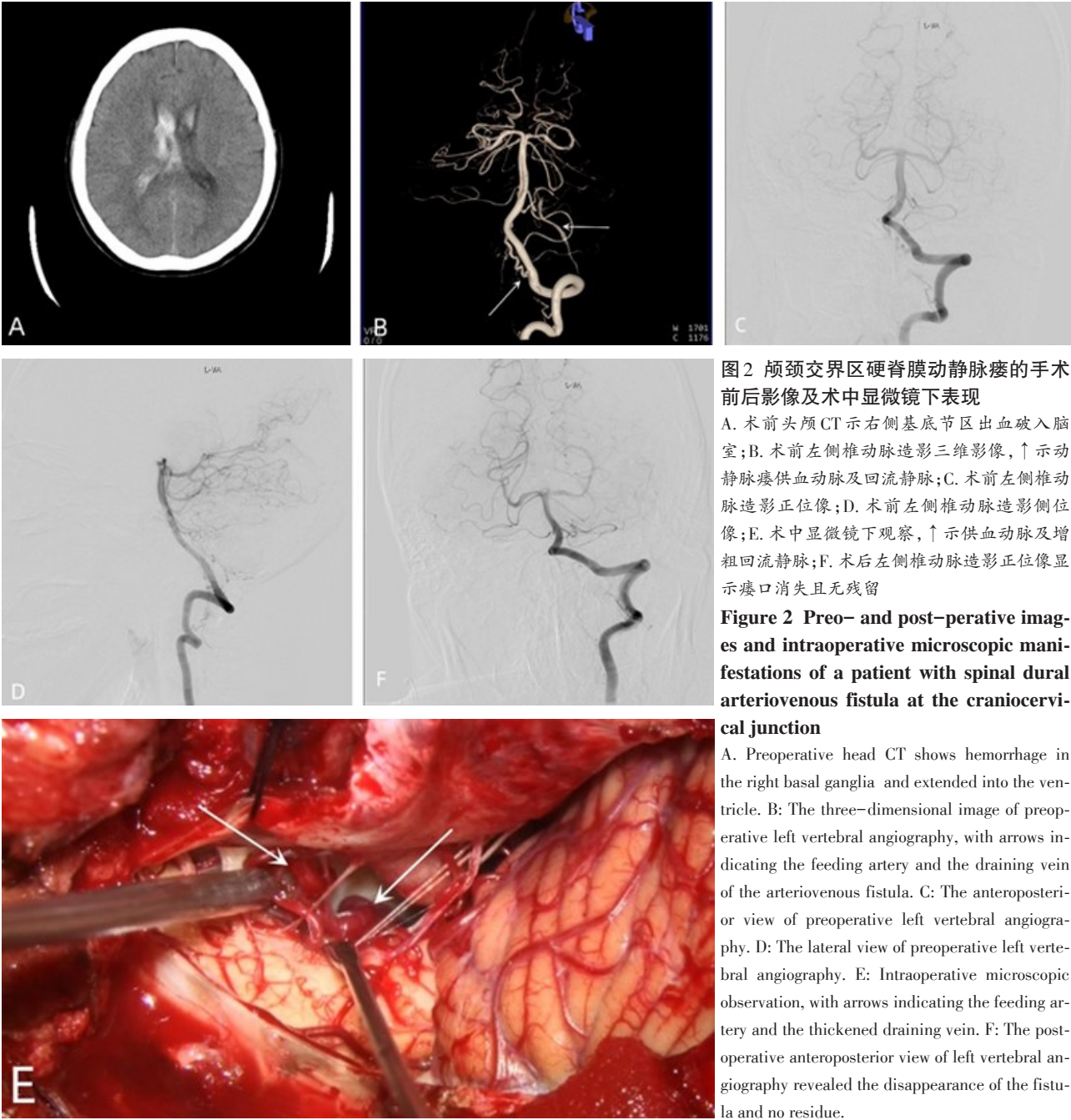


图2 颅颈交界区硬脊膜动静脉瘘的手术前后影像及术中显微镜下表现

A. 术前头颅CT示右侧基底节区出血破入脑室;B. 术前左侧椎动脉造影三维影像,↑示动静脉瘘供血动脉及回流静脉;C. 术前左侧椎动脉造影正位像;D. 术前左侧椎动脉造影侧位像;E. 术中显微镜下观察,↑示供血动脉及增粗回流静脉;F. 术后左侧椎动脉造影正位像显示瘘口消失且无残留

Figure 2 Pre- and post-operative images and intraoperative microscopic manifestations of a patient with spinal dural arteriovenous fistula at the craniocervical junction

A. Preoperative head CT shows hemorrhage in the right basal ganglia and extended into the ventricle. B: The three-dimensional image of preoperative left vertebral angiography, with arrows indicating the feeding artery and the draining vein of the arteriovenous fistula. C: The anteroposterior view of preoperative left vertebral angiography. D: The lateral view of preoperative left vertebral angiography. E: Intraoperative microscopic observation, with arrows indicating the feeding artery and the thickened draining vein. F: The postoperative anteroposterior view of left vertebral angiography revealed the disappearance of the fistula and no residue.

预后与病程长短、术前脑脊髓功能状态等相关,因此提高对该疾病的认识,早期明确诊断是提高疗效的关键,且须及早手术治疗,闭塞瘘口,改善正常脑脊髓血流灌注。

总之,CCJ-SDAVF是一少见脊髓血管病,选择性脑脊髓血管造影是诊断的金标准,应早发现、早治疗,显微手术或血管内介入栓塞治疗为主要治疗方案。显微手术灼闭瘘口是治疗颅颈交界区硬脊膜动静脉瘘的有效治疗方案。

署知情同意书。本研究方案于2024年3月11日经中国人民解放军中部战区总医院医学伦理委员会审批,批号为:202406801。

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(上接第 659 页)

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